

ABSTRACT

Background:

Hemophilia is a rare inherited X-linked recessive disorder characterized by deficiency of Factor VIII or Factor IX coagulant activity required for generation of Thrombin in the Intrinsic pathway of coagulation. Hemarthrosis accounts for ~90% of bleeding in hemophilic patients. With increase in severity of the disease, patients experience increased frequency of spontaneous bleed, which further leads to irreversible joint destruction. The aim of the study is to evaluate hemophilic joints clinically, functionally and radiologically.

Materials and Methods:

Our study population were 60 patients (4-18 years), confirmed Factor VIII/IX deficiency attending Government Royapettah Hospital. We further classify them as having Mild (6-40%), Moderate (1-5%), Severe (<1%) according to their factor activity level. All patients were subjected to thorough history taking and examination and X-ray of target joints. Target joints were evaluated using Hemophilia Joint Health Score 2.1(HJHS), Functional Independence Score in Hemophilia (FISH) and Pettersson scoring.

Results:

Patients with severe hemophilia comprises 70% of study population. Ankle was the most common target joint. The age at hemarthrosis and average bleed per year

showed statistically significant correlation with severity of disease. HJHS and Pettersson score were significantly higher in those with severe disease, while on the other hand, FISH was significantly lower. There was also a significant correlation of the 3 scoring system with frequency of joint bleed.

Conclusion:

There was significant impairment of joint in people with severe hemophilia. HJHS, FISH and Pettersson score proved to be useful tools for assessment of joint health in Hemophilia.

Keywords:

Severe hemophilia, Hemarthrosis, Hemophilia Joint Health Score, Functional Independence Score in Hemophilia, Pettersson Score